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ABSTRACT

This guide provides information on Batten Disease to assist in planning a quality educational program for the student with the disease. Because Batten Disease, or neuronal ceroid lipofuscinosis, causes the death of brain cells, students with the disease are described as suffering from mental impairment, worsening seizures, and progressive loss of sight and motor skills. Eventually, these children become blind, bedridden, and unable communicate. The disease is always fatal, typically by the late teens or twenties. The guide discusses the following: (1) history of the condition; (2) types of the disease; (3) etiology; (4) diagnosis; (5) characteristics of students; (6) visual impairments and suggested interventions; (7) muscular control and strategies for supporting fine and gross motor skills; (8) physical therapy and Batten Disease; (9) social interaction; (10) cognitive impairments and classroom strategies; (11) speech/language impairments and suggestions for promoting speech; (12) communication and using daily calendar boxes to enable the student to plan their day; (13) Individualized Education Program goals; and (14) student rights under the Individuals with Disabilities Education Act of 1997. The guide closes with some notes for the school nurse. (CR)

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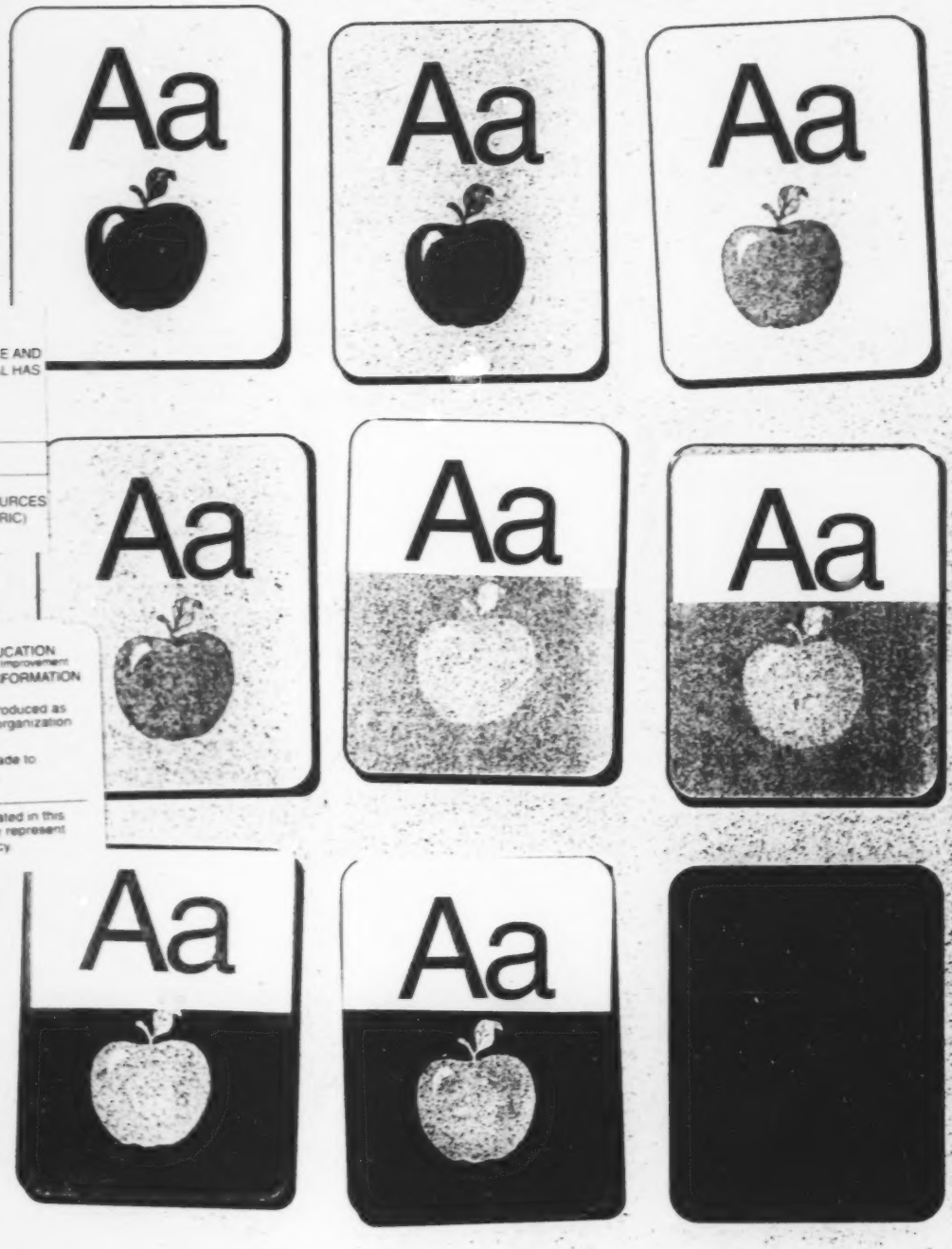

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Teach and Be Taught

A Guide to Teaching Students with Batten Disease

A challenge awaits you. That challenge is to educate your student who has Batten Disease. This guide will help you improve the quality of life for your student and assist the family in accepting the challenges that they face. Batten Disease offers little hope at this time in the medical arena. However, the progression of the disease is predictable. This guide, utilizing the predictability of the disease provides the knowledge base to planning a quality educational program for the student with Batten Disease. It is vital to know the progression and end results of the disease to make time count concerning education for the student with Batten Disease. What you teach now can benefit and empower the student even in the final stages of the disease. Recognize, though, that there will be individual differences among students, requiring education to be individualized according to students' individual needs.

What is Batten Disease?

Batten Disease is named after the British pediatrician who first described it in 1903. The juvenile form is the most common form of a group of disorders called neuronal ceroid lipofuscinosis (or NCLs). Although Batten Disease is usually regarded as the juvenile form of NCL, it has now become the term to describe all förms of NCL. The basic cause, progression, and the outcome are the same. The forms of NCL are classified by age of onset but are all genetically different.

Over time, affected children suffer mental impairment, worsening seizures, and progressive loss of sight and motor skills. Eventually, children with Batten Disease become blind, bedridden, and unable to communicate. Batten Disease is always fatal typically by the late teens or twenties. Batten Disease is not contagious or, at this time, preventable.

History of Neuronal Ceroid Lipofuscinosis

The first probable instances of this condition were reported in 1826 by Dr. Christian Stengel in a Norwegian medical journal, who described 4 affected siblings in an small mining community in Norway. Although no pathological studies were performed on these children the clinical descriptions are so succinct that the diagnosis of the Spielmeyer-Sjogren (juvenile) type is fully justified.

More fundamental observations were reported by F. E. Batten in 1903, and by Vogt in 1905, who performed extensive clinicopathological studies on several families. Retrospectively, these papers disclose that the authors grouped together different types of the syndrome.

In 1913-14 M. Bielschowsky delineated the Late Infantile form of NCL. However, all forms were still thought to belong in the group of "familial amaurotic idiocies", of which, Tay-Sachs was the prototype. Subsequently, it was shown by Santavuori and Haltia that an infantile form of NCL exists, which Zeman and Dyken had included with the Jansky-Bielschowsky type.

What are the forms of NCL/Batten Disease?

There are four main types of NCL, including two forms that begin earlier in childhood and a very rare form that strikes adults. The symptoms are similar but they become apparent at different ages and progress at different rates.

Infantile NCL (Santavuori-Haltia disease): begins between about 6 months and 2 years of age and progresses rapidly. Affected children fail to thrive and have abnormally small heads (microcephaly). Also typical are short, sharp muscle contractions called myoclonic jerks. Initial signs of this disorder include delayed psychomotor development with progressive deterioration, other motor disorders, or seizures. The infantile form has the most rapid progression and children live into their mid childhood years.

Late Infantile NCL (Jansky-Bielschowsky disease) begins between ages 2 and 4. The typical early signs are loss of muscle coordination (ataxia) and seizures along with progressive mental deterioration. This form progresses rapidly and ends in death between ages 8 and 12.

Juvenile NCL (Batten Disease) begins between the ages of 5 and 8 years of age. The typical early signs are progressive vision loss, seizures, ataxia or clumsiness. This form progresses less rapidly and ends in death in the late teens or early 20s, although some may live into their 30s.

Adult NCL (Kufs Disease or Parry's Disease) generally begins before the age of 40, causes milder symptoms that progress slowly, and does not cause blindness. Although age of death is variable among affected individuals, this form does shorten life expectancy.

How many people have these disorders?

Batten Disease and other forms of NCL are relatively rare, occurring in an estimated 2 to 4 of every 100,000 births in the United States. The disease has been identified worldwide. Although NCLs are classified as rare diseases, they often strike more than one person in families that carry the defective gene.

What causes these diseases?

Symptoms of Batten Disease and other NCLs are linked to a buildup of substances called lipopigments in the body's tissues. These lipopigments are made up of fats and proteins. Their name comes from the technical word lipo, which is short for "lipid" or fat, and from the term pigment, used because they take on a greenish-yellow color when viewed under an ultraviolet light microscope. The lipopigments build up in cells of the brain and the eye as well as in skin, muscle, and many other tissues. Inside the cells, these pigments form deposits with distinctive shapes that can be seen under an electron microscope. Some look like half-moons (or commas) and are called curvilinear bodies, others look like fingerprints and are called fingerprint inclusion bodies and still others resemble gravel (or sand) and are called granular osmiophilic deposits (GRODS). These deposits are what doctors look for when they examine a skin sample to diagnose Batten Disease.

The biochemical defects causing NCLs have not been identified. Some scientists suspect these abnormal deposits result from a shortage of enzymes normally responsible for the breakdown of lipopigments. According to this theory, diseased cells produce inadequate amounts of enzymes or manufacture defective enzymes that function poorly. As a result, the cells cannot process enough of the lipopigments that occur within them, and the lipopigments accumulate. However, scientists have not pinpointed what specific enzymes are at fault or determined how the stored lipopigments damage nerve cells.

Other scientists believe that abnormal lipopigment buildup may result from a glitch in the cell's production or processing. For example, diseased cells could be producing too much of a normally needed lipoprotein.

How are these disorders diagnosed?

Because vision loss is often an early sign, Batten Disease may be first suspected during an eye exam. An eye doctor can detect a loss of cells within the eye that occurs in the three childhood forms of NCL. However, because such cell loss occurs in other eye diseases, the disorder cannot be diagnosed by this sign alone. Often an eye specialist or other physician who suspects NCL may refer the child to a neurologist, a doctor who specializes in disease of the brain and nervous system. In order to diagnose NCL, the neurologist needs the patient's medical history and information from various laboratory tests. Diagnostic tests used for NCLs include:

Blood or urine tests:	These tests can detect abnormalities that may indicate Batten Disease. For example, elevated levels of a chemical called dolichol are found in the urine of many NCL patients.
Skin or tissue sampling:	The doctor can examine a small piece of tissue under an electron microscope. The powerful magnification of the microscope helps the doctor spot typical NCL deposits. These deposits are common in skin cells, especially those from sweat glands.

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Electroencephalogram/EEG: An EEG uses special patches placed on the scalp to record electrical currents inside the brain. This helps doctors see telltale patterns in the brain's electrical activity that suggest a patient has seizures.

Electrical studies of the eyes: These tests, which include visual-evoked responses (VER) and electro-retinagrams (ERG), can detect various eye problems common in childhood NCLs.

Brain scans: Imaging can help doctors look for changes in the brain's appearance. The most commonly used imaging technique is computed tomography (CT), which uses x-rays and a computer to create a sophisticated picture of the brain's tissues and structures. A CT scan may reveal brain areas that are decaying in NCL patients. A second imaging technique that is increasingly common is magnetic resonance imaging, or MRI. MRI uses a combination of magnetic fields and radio waves, instead of radiation, to create a picture of the brain.

Is there any treatment?

As yet, no specific treatment is known that can halt or reverse the symptoms of Batten Disease or other NCLs. However, seizures can sometimes be reduced or controlled with anticonvulsant drugs, and other medical problems can be treated appropriately as they arise. At the same time, physical and occupational therapy may help patients retain function as long as possible.

Some reports have described a slowing of the disease in children with Batten Disease who were treated with vitamins C and E and with diets low in vitamin A. However, these treatments did not prevent the fatal outcome of the disease.

Support and encouragement can help children and families cope with the profound disability and losses caused by NCLs. Meanwhile, scientists pursue medical research that could someday yield an effective treatment.

What research is being done?

Within the Federal Government, the focal point for research on Batten Disease and other neurogenetic disorders is the National Institute of Neurological Disorders and Stroke (NINDS). The NINDS, a part of the National Institutes of Health (NIH), is responsible for supporting and conducting research on the brain and central nervous system. The Batten Disease Support and Research Association and the Children's Brain Diseases Foundation also provide financial assistance for research.

Through the work of several scientific teams, the search for the genetic cause of NCLs is gathering speed.

In September 1995, The International Batten Disease Consortium announced the identification of the gene for the juvenile form of Batten Disease. The specific gene, CLN3, located on Chromosome 16, has a deletion or piece missing. This gene accounts for 73% of all cases of Juvenile Batten Disease. The remainder are the result of other defects of the same gene.

Also, in 1995, scientists in Finland announced the identification of the gene responsible for the infantile form of Batten Disease. The gene, CLN1, is located on Chromosome 1.

In September 1997, scientists at the Robert Woods Johnson Medical School and the Institute for Basic Research, NY, announced the identification of the gene for the "classic" late infantile form of Batten Disease. The gene, CLN2, is located on chromosome 11.

Research continues to attempt to identify the genes for a Finnish form and variant form of late infantile, the genes for which appear to reside on chromosomes 13 and 15 respectively. Research continues toward identification of the gene for the adult form of Batten Disease, also known as Kufs Disease.

Identification of the specific genes for Infantile, Late Infantile, and Juvenile Batten Disease has led to the development of DNA diagnostics, carrier and prenatal tests.

What is happening to the student ?

Batten Disease is causing death of neurons (brain cells). As the neurons die more and more symptoms of the disease become apparent. Seizures begin and continue to intensify as time passes. The child may suffer from many different types of seizures, typically absence (petit mal), tonic clonic (grand mal), atonic (drop), myoclonic (sudden jerks) and complex partial (psychomotor) are seen with Batten Disease. However, one key to understanding Batten Disease is to understand that the illness knows no specific pattern or time schedule. One child may have one type of seizure and another may have two different types. One child may have an onset of seizures at age five another at age 9 and still another at age 13. The teacher and staff need to have an understanding of what seizures are and what to do if a child has a seizure. One thing is for certain; DO NOT PLACE ANYTHING IN THE CHILD'S MOUTH DURING A SEIZURE !

As was previously noted, one of the initial symptoms of Batten Disease is the beginning loss of vision. The vision loss, once started, is not reversible and will lead to total blindness. During the course of the vision loss the child will become color blind and will loose central vision first. You may see the child eventually looking at objects, people, etc. out of the corner of the eye. This peripheral vision will usually last for awhile. When the peripheral vision is lost the child will be left with light/dark perception. There have been many reports from children and their parents that before total blindness occurs there is a period when the totality of darkness will come and go.

An additional symptom of Batten Disease is "clumsiness". The disease affects the parts of the brain that deal with mobility and clumsiness is evident soon after or during the onset of the illness. As the illness progress the ataxia worsens and mobility will continue to decline. Batten Disease causes eventual total loss of mobility.

In addition to seizures, blindness and mobility loss, as the brain cells die and the disease progresses there will be other losses such as: loss of the ability to eat, loss of speech, loss of continence, and loss of cognizance.

There are two things to remember. The first is that there is no timetable when these things will happen. We can say in general that these things will occur but not when or even in what order they will happen. The second is that all of this is slow and will happen over many years. Batten Disease has been compared to the teeth of a circular saw. There are drops when the child will suffer a loss or losses, a rebound and then a plateau or level time. No one can predict when a drop or loss will occur and what may precipitate it. No one can say how much of a rebound there will be. The plateaus will usually last for some time, perhaps weeks and even months before another drop or loss is experienced. The plateaus are welcome and valuable periods in that they allow the child, family and teacher time to adjust.

These children in early years will attain developmental goals on or even ahead of schedule. Things such as crawling, sitting, walking, word development, numbers, shoe tying, talking, sentence development, and so on, right into math, language, and writing are normal with these children.

If the learning curve of a child with Batten Disease was compared to a "normal" child one would see that they are alike until after the onset of the illness. After onset, if the learning curve could be charted, one would see it begin to slow in comparison. As time advances the curve will begin to flatten and the time will come when the child will "peak".

When a child with Batten Disease begins to peak it is common to see some areas of learning begin to stop and others continue to advance, although slowly. Many reports have been made of students who can perform tasks and later in the same day not be able to perform the same tasks and still later be able to accomplish it properly. It is when this peak is reached that a teacher needs to consider a reverse trend in thinking that will correspond to the reversal that the child will undergo. Until the peak is reached the child should be challenged to continue to learn, the difference being that expectations need not be as high as would be for a child that will not experience neurologic regression.

When the child has passed beyond the peak the major regression will begin. The downhill decline begins and does not end. The child loses things in a reverse order. In other words, the last thing learned will be the first loss. One will observe that as the regression continues the child will lose short term memory and retain long term memory for a long time.

It is when the regression begins that the teacher becomes challenged. Teachers are taught to teach. How do you deal with "unlearning" ? The following is an illustration how one teacher solved the problem. A girl with Batten Disease loved spelling. She began with the usual easy words, i.e. dog, cat, etc. and progressed to where she was spelling large difficult words. When she began to peak she would be able to correctly spell her list of words and a short time later not be able to spell any of them. Given an hour or two break and she would once again correctly spell all the words but again later not be able to spell them. It was back and forth. Once she really began to regress the teacher adjusted her word list by going backwards and giving her easier words. She eventually ended up back at the simple words of dog and cat. The teacher heaped praise on her and she in turn glowed in the praise and felt a sense of accomplishment.

Vision Information for the Regular Education Classroom Teacher

With this progressive degenerative condition, loss of vision will occur. At first the visual loss may not be apparent but the signs of visual loss will increase in the classroom setting. The teacher needs to be aware of the following symptoms:

1. Changes in visual acuity.
2. Inconsistent and erratic performance of far point visual tasks such as seeing the writing on the chalkboard or a poster from the student's desk across the room.
3. Squinting and shifting to focus on near point visual tasks such as reading at the student's seat or while trying to assemble a project directly in front of the student.
4. Asking more frequent questions or asking for verbal repetition of instructions regarding the student's own performance when vision is part of the learning activity.
5. Gradual shift in participation in classroom activities that require vision.
6. Changes in physical proximity to the sources of instruction or activities. The student may seek out a closer location initially and later may not realize to move closer to those sources for information.

Among the interventions that the classroom teacher should consider for the student with Batten Disease include:

1. Teacher and speaker in close proximity to the student.
2. Preferential seating for activities where usable vision will enhance learning such as sitting closer to the speaker or nearer the chalkboard.
3. Providing more light at the student's work areas to promote the use of residual vision. Intensify the light source. Mix the types of lighting, trying to include natural light as well as florescent and incandescent lighting.
4. Give clear large or enlarged print examples and orally discuss those examples, asking the student questions which will reflect the learner's understanding of the presented concepts and ideas.
5. Use clearly contrasting materials in visual presentations. A dark or bold ink on an off white or cream colored paper will stand out more than the pastel shades of colored pencils or papers. Ask the student which inks and paper stand out or work best.

6. Try to use original dark ink material rather than purple dittoes or penciled work for the student's ease in reading.
7. Show graphs, charts, diagrams, pictures, and other visual models to the student directly with verbal discussion and accompanying comprehension questions.
8. When using typed print, choose a good quality print that is legible to the reader.
9. When using computer generated materials, select laser prepared print over dot matrix print for clarity.
10. Place a yellow transparency film over purple colored materials to intensify the legibility.
11. Assign a "buddy" and have the students work in cooperative pairs, repeating the instructions auditorially to each other and especially to the student with Batten Disease.
12. Start to increase the amount of oral as well as written directions to tasks asked of students in your classroom.
13. Have the students examine three dimensional learning materials more frequently in the classroom such as a relief map of their state, feeling landforms on a world globe, or studying time by manipulating unifex cubes to represent minutes on a large clock face.
14. Ask the student to be part of demonstrations and part of role playing that is done in the classroom to encourage more active participation as long as possible.
15. Enlarge the print on presented materials, even if this means using two pages for presenting the content of a single page.
16. Bring in more manipulative objects for the student to study. Relate these directly to the learning tasks at hand.
17. When new items are posted in the room, make a point to tell and take the student to these locations (i.e. a bulletin board, the door, on the wall, or at a table) so he/she can learn the new information.
18. Present a variety of writing instruments to the student and let him or her to select the one(s) that he/she can use with more legibility.
19. Reinforce with verbal praise and positive comments to the student when he or she attempts to learn actively in the classroom.
20. Incorporate the student's known information into the beginning discussion of unknown information.
21. Use a print enlarger when visual documents need enlarging.
22. Ask the vision specialist for other specific suggestions which may be appropriate for the student in your classroom setting.

Discuss with the parents how well the student handles a variety of visual tasks in the classroom. Indicate the successful as well as the less successful situations. Inquire of the parents if they have found any particularly effective techniques to deal with the visual loss in the home that might be tried in your classroom also. As the student's vision will continue to change, ask the parents to update you on any visits to their vision specialist or contact with medical personnel regarding their son or daughter's condition. Above all, remember this is a team effort of parents and professionals where sharing and communicating needs to take place much more frequently than with other students and their families. Remind them that you are there to be part of the support team to help their son or daughter. Your care is valued and appreciated by the families.

Muscular Control

The area of muscular control encompasses both fine and gross motor abilities. Fine motor skills include handwriting, dressing, block building, and many other skills that require finger dexterity and coordination. Gross motor skills include walking, running, playing sports, and many other skills that use the large muscle groups of the arms and legs. Muscular control is a focus of the educational process due to the regression of these skills as the disease progresses.

Fine Motor. Fine motor activities become increasingly difficult for the student with Batten Disease. The student knows what he or she wants to do but often can not make the fingers perform. This periodic breakdown between perception of the brain and the actual motor act causes frustration that results in the student acting out inappropriately in many cases. Therefore, it is the teacher's responsibility to read the student and recognize the level at which to challenge and when to simply nurture. For example, a particular young student loved to build with Lego blocks. He built intricate structures and was very proud of his creations. However, as his abilities declined, he engaged in this activity less and less. A very wise teacher recognized what was happening and adapted the student's block building activity by providing bigger blocks that were easier to put together. The student once again happily engages in his favorite activity and creates elaborate, bigger, structures. This teacher read her student and recognized how and when to challenge him.

Braille, a skill often taught to students with visual impairments, becomes more of a fine motor difficulty than a cognitive task for the student with Batten Disease. The numbness that occurs in the fingers of such students impairs their ability to interpret the small raised dots. The philosophy in Holland concerning Braille is to teach it if the student shows an interest in learning this form of communication. Otherwise, if no interest is shown, don't waste time trying to teach Braille. This skill will diminish early, and time can be more wisely spent teaching skills that will endure the disease process.

Ultimately, read your student. Focus on the fine motor activities that he or she enjoys and adapt those activities to allow the student to be successful.

Gross Motor. Gross motor activities center around mobility, endurance, and strength. Two very different schools of thought have arisen in the area of gross motor skills. One, provide massive amounts of therapy to maintain gross motor abilities for as long as possible. On the other hand, the school of thought from Holland is that it is sadistic to prolong something that is slipping away. Both philosophies show merit. Therefore, this decision becomes a personal choice of the family. However, when the physical therapy becomes very stressful to the student it is time to back away from that activity. Undue stress may increase seizure activity. This trade-off is not worth pursuing.

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Again, read your student when balancing activities that address gross motor skills. For example, one young girl loved her gymnastics class that she attended once a week. She also received physical therapy twice a week. As therapy became too stressful, the mother decided to decrease physical therapy to once a week and increase gymnastics class to twice a week. A student that is losing his/her ability to walk may find enjoyment by practicing walking in a swimming pool. The buoyancy of the water enables the student to continue to walk when the ability out of the pool is no longer possible. The swimming pool will also stimulate circulation and allow the student a better sense of where his/her body is in space.

Physical Therapy and Batten Disease

Normal Motor Development

It has been said, ***"We learn to move and move to learn".*** Normal motor development, or more specifically the acquisition of gross motor skills, is observed through the develop- ment of 'motor milestones' (e.g., head-up, roll, sit-up, creep, stand, & walk). These motor milestones along with the brain's developing ability to organize and process sensory stimuli, enable an individual to respond, move about, orient, and function in various and changing environments.

The body's motor responses to environmental changes are known as 'postural' and 'equilibrium reactions' and are observed in adjustments made in posture, protective reactions, balance, and equilibrium. Physical therapists are able to measure the quality of these movements and make adjustments to provide information regarding the individual's safety, motor dependence or independence, functional/developmental level, motor progression/regression status, and intervention strategies.

The motor system does not develop in isolation. Closely integrated is the sensory system which organizes sensations that in turn elicit a motor response. This sensory-motor system in the early years is the foundation for more complex skill functions such as writing, reading, behavior and social skills. As a child with Batten Disease develops, many sensorimotor skills will be affected, some skills will be lost and others need be relearned in an adapted way.

Implications of Batten Disease

Important considerations must be noted in dealing with children with Batten Disease. The progressive regression and devastating effects of Batten Disease is not always predictable; the intensity and timetable of physical decline and involvement is unpredictable. Unknown individual factors are the order and degree of involvement and losses and each child's tolerance level to these losses.

As with other diseases or injury to the brain, symptomology and ramifications of Batten Disease can be widespread and extensive. Deterioration is documented in motor, sensory, communicative, behavioral, cognitive and psychosocial functioning. This is often observed in vagueness or declines in self-awareness, problem solving, judgment, attention and information processing, including short-term memory and thought organization.

Physical and motor symptomology

Because of the marked deteriorating nature of Batten Disease, thorough evaluations, reassessments, accurate and measurable record keeping and documentation are highly important.

The motor disorders observed with Batten Disease at first may be subtle and inconsistent. However, as the disease progresses, symptomology increases as does the intensity of the disorders. Deterioration may be evidenced in several of the following:

1. Diminished postural mechanism (loss of head righting, protective reactions)
2. Loss of balance/equilibrium (clumsiness, stumbling, falling)
3. Deteriorating posture (bowed-rigid, linear, lacks rotation, increased flexion of extremities, knees bent, 'sinks' into gravity)
4. Changes in gait pattern (wider base, toeing-in, shuffle gait, crouched-gravitational insecurity)
5. Changes in breathing patterns (shallow, short)
6. Abnormal muscle tone
   - hypertonic (usually extremities-spastic/rigid lacks full range-of-motion)
   - hypotonic (usually of trunk with decreased stability)
7. Ataxia (lack of graded control of movement of trunk and extremities)
8. Extraneous-involuntary movements
9. Muscle weakness and atrophy
10. Decreased facial expression (mask like)
11. Difficulty initiating actions (swallowing, speech, transfers)
12. Tremors (often combined with ataxia and spasticity; limits gross & fine motor)
13. Apraxia (difficulty in motor planning, affects fine/gross/oral motor tasks/swallow)
14. Dysarthria (collection of oral motor disorders-speech, chewing, swallowing)
15. Decreased motor planning, orientation, localization, language-word retrieval

Physical therapy interventions

The role of physical therapy in the school system is evaluating, identifying, and developing motor goals that promote and facilitate maintenance of gross motor skills, maximize the quality of movement and function, and provide support through adaptive equipment when necessary. The ultimate goal is to enhance the student's well being and continued learning, even if through different modes and at different paces.

The following recommendations are not prioritized in order of importance or need. These are only a few suggestions for therapy and many times activities will yield benefits for more than one area. Again, the underlying goal is to enhance the child's well-being and quality of life while working within his/her ability and tolerance levels.

Maintaining gross motor skills while maximizing function

1. Provide opportunities for mobility. (ball, pool, or mat exercises instead of standing).
2. Provide opportunities for deep and increased breathing.
3. Motivate with enjoyable activities. (combine motor movements with auditory stimuli e.g., stationary bike-exercise to music).
4. Maintain touch as tolerated. (use firm contact & avoid light touch if not tolerated).
5. Maintain proper body alignment, especially in sitting and laying.
6. Continue standing and weight bearing. Use adaptive equipment as needed.
7. Provide a peaceful, safe, positive, and non-threatening environment.

Provide support and security

1. Provide safety bars and physical support for transfers and ambulation.
2. Utilize splints and braces as needed to enable weight bearing and mobility.
3. Utilize adaptive equipment to provide secure, well aligned sitting, standing and ambulation.
4. Support wheelchair use for ease of travel for child and family as needed.
5. Provide recommendations for "in-home" physical management.
6. Always provide moral support to student and family.

Prevent injuries and deformities

1. Plan ahead! Allow additional time and avoid hurrying for all activities.
2. Provide a familiar, secure, barrier free environment for safety & ease of exploring.
3. Facilitate mobility & weight shifting as long as possible (e.g., bed mobility, reaching, come-to-sit, roll, etc.)
4. Adapt activities of daily living as needed. (foods easy to chew & swallow, straw for ease in drinking, tub/shower seat with spray hose, elevated toilet seat, safety bars for all transfers, bed-rail, avoid slippers/throw rugs)
5. Insure a well-aligned posture in positions.

The importance of active movement and mobility cannot be over emphasized!
It has been researched and reported that sensations and the motor responses to sensations act as nourishment to the brain. However, due to each child's individual tolerance level and abilities, the role of the educator is to encourage, facilitate and assist in active mobility as long as it is possible and enjoyable for the child. When active participation becomes stressful, intervention strategies need to be changed with more emphasis on comfort, support, assistance and adaptive equipment.

A full team approach with professionals, parents and the student working together will accomplish the most in promoting the greatest potential for quality-of-life for the child with Batten Disease.

Social Interaction

Social interaction becomes the biggest issue when questions arise concerning the educational placement. Many families have a difficult time removing their child from a regular education setting to a more restrictive educational placement. However, do not allow the inclusion panacea to sacrifice the well being of the student. Remember, students with Batten Disease know what is happening to them. They know what they once could do that their peers are still able to do. This knowledge often surfaces as problematic behaviors. The student may be trying to tell those around him or her that it is extremely painful to be educated with peers, with whom it is impossible to keep up with. Perhaps, this is the time to place the student in a more restrictive educational placement. Social interaction with familiar peers can continue but on a different level, such as peer tutoring or mainstreaming for subjects that the student still enjoys participating in with his or her nondisabled peers. Once again, it is vital to read the student.

The child wants to maintain independence. However, as the illness progresses the child has to be moved towards dependence. Teaching tactics will eventually change from teaching new things to teaching and maintaining daily living skills.

Cognition

Dementia is defined as the loss of cognizance. Batten Disease can bring a whole new meaning to the word. For some children the dementia process can be extreme in that it can produce severe "behavioral problems". These may range from severe outbursts of anger, uncontrollable emotions, striking out, hitting, biting, hysterical laughter, heart wrenching sobbing and hallucinations. Like many other aspects of Batten Disease how the dementia process may affect a child is not predictable. Some children have no problems and others have severe problems. The remainder are somewhere in between. The dementia process is also the least understood and most difficult with which to deal.

In the moderate to severe form the dementia can become almost intolerable. The uncontrolled behavior is at best an almost impossible situation, especially if the child becomes vocal. *Echolalia* is repetitive speaking. The child may ask a question over and over and over and although you may give an answer, the child continues to ask it again and again. Usually, this period is relatively short lived and will pass. The often most difficult time is when the child's thought process becomes disrupted. The outbursts can come at any time without warning and usually for no apparent reason. No amount of logical resolution will work to deter what the child may have on his/her mind at the time. It is especially important to be aware that these children still understand the difference between appropriate and inappropriate behavior. They often cannot help what is happening. Hallucinations usually seem to take on two aspects. Either persons who are obviously not present or terrible things such as snakes, spiders and monsters. We know that all of this is not real but to the children they are very real and threatening.

All of this can be disruptive to the classroom setting. You, the teacher, and the other classmates need to understand that the child is usually not able to control what is happening. Understanding and empathy are needed. Yet the social interaction with the child's peers is important. There is no good resolution to this to this problem. Often times it is trial and error as to what will work best to bring quiet and order to the classroom. Usual behavior modification techniques may be ineffective.

There are a couple of things that is known to help. One is that as the dementia process takes hold the child does better on a strict regimented schedule. It would seem that it is something that they are able to hang onto and understand. They seem to have difficulties handling changes to routine or normalcy, especially sudden unexpected changes. When the dementia is severe you may see that anticipation and disappointment can cause problems also. An example would be that the class will go on a field trip in four days to the zoo. During the intervening time the anticipation of this event may build and build to a bursting point. If, for some reason the field trip is canceled you will probably have difficulty with the child having Batten Disease as the reason for the cancellation will not be understood. The anticipation alone may cause problems just because it is anticipation that cannot be controlled. If you know that anticipation and/or disappointment are becoming a problem consider not telling the child until about time for the event to happen. This may appear contridictive to the previous statement relative to sudden changes to schedule. Changes that lead to things that are pleasurable to a child may not cause any difficulty. You may have to weigh one against the other.

The other thing that is often helpful is playing a child's favorite music or video tape during an outburst or when there is difficulty. We know that these children have favorite music or videos that they seem to hang onto and will listen to over and over. Be assured that the period of dementia will pass. No one can predict how long it will last, but what is usually noted is a gradual abating of the dementia until it is completely gone. Unfortunately it also signals the end of many other functions.

Several drugs have been tried to ease the dementia problems the children experience. Sometimes they help, sometimes they help for awhile, and sometimes they have an opposite affect to that desired and sometimes the drugs have no affect at all. Like everything else with Batten Disease there is no single drug or method that will work for every child.

A key point in all of this is that the child is aware of what is happening to him/her. He/she is aware of the continuing slowing in abilities and eventually the losses that take place cognitively, emotionally, and physically. This awareness can exacerbate the dementia. Frustration levels can be exceeding high for the child when he/she cannot retrieve words, move about as easily, use hands and fingers and express him/herself as well as before.

The dementia that occurs with Batten Disease appears somewhat different from other neurologic regressive diseases. Long-term memory stays quite intact. This fact is very significant to you as the teacher of a student with Batten Disease. Focus instruction on areas that will be important and meaningful to the student in the future. For example, teach object cues that correspond with activities that the student enjoys even when the student still has verbal communication skills.

Objects taught early are more likely to remain in long-term memory and will allow the student to access them when the verbal skills are gone. This area is covered in depth in the section on communication.

Students with Batten Disease love to reminisce about their past. Discuss past events with them often to help secure significant events in their long-term memory. These memories bring comfort and stability to a life that is facing chaos.

The most important aspect of cognition is for the teachers and caregivers to realize that the student with Batten Disease possesses a great deal of intelligence even when he or she cannot access this knowledge. Never underestimate this intelligence. Formal assessment suggests a decrease in IQ However, this decrease may be due only to a decrease in performance, rather than an actual decrease in IQ What these students actually know and understand is unknown and untestable. Regardless, assume that your student knows and understands everything and never talk down to him or her. Always treat your student with the same dignity and respect that a normal peer enjoys. Emily, a fifteen-year-old girl with Batten Disease, would become extremely agitated when her teacher did not understand this and would sing nursery rhymes to comfort her. This only enraged Emily further.

Once the teacher understood, she was able to make progress with Emily. They were often found in discussions about boys, philosophy, friends, family, and even death. These discussions pleased Emily, calmed her, and stimulated further intellectual thoughts. Emily would smile rather than cry. The agitation was gone.

In Holland, the day for students with Batten Disease begins with the teacher reading the current events from the newspaper. The students appear happy and content as they intently listen to what is happening in the world around them. The written word brings back memories and comforts the student with Batten Disease. Trevor is a fourteen-year-old boy with Batten Disease. Trevor has little control over his jerky movements due to the seizure activity and neurologic damage resulting from Batten Disease. However, he loves listening to books on tape, and he will work extremely hard at controlling his excess movements when activating the tape recorder with a switch (fig.1). Even in the advanced stage of the disease Trevor knows that releasing the pressure on the switch will result in the reader stopping as well. Favorite books from Trevor's past bring him great joy as he actively and alertly accesses them in this way.

Figure 1. Trevor activates his book on tape with a switch. Trevor works hard to maintain pressure on the switch, so his favorite stories continue to play.

Speech/Language

The following is a list of characteristics found in the early stages of Batten Disease from a speech/language perspective. A diagnosis of retinitis pigmentosa in a young child should raise a red flag for professionals working with the child. This child should be monitored closely. Watch for:

- a plateau of academic skills
- hesitations, repetitions, and stuttering-like speech
- a somewhat awkward gait, with the knee or foot turned slightly inward
- blank stares that may only last a few seconds

The Speech/Language suggestions that follow may prove beneficial with a child in the early stages of Batten Disease.

1. Early on, one can tell that the child's ability to comprehend language supersedes their ability to express language. Thus, one may find ways to help this child express his/her thoughts; verbally, with sign language, and/or with augmentative communication devices. As the disease progresses, one must always remember to provide input.

2. Due to the progressive visual loss that the child experiences, professionals will need to direct their input to the auditory, tactile and motor modalities. Using these modalities, it is imperative that the teacher keep the child actively engaged in the activity being addressed.

3. Gain the child's attention before giving oral information, i.e. "Show me you are listening. Turn on your ears." etc. In addition, addressing the child by name may also get his/her attention.

4. Listening can be a passive activity. Incorporate the use of objects to elicit speech when you want the child to retell stories, answer questions, recite poems, and sequence rote information. When using this technique, the child's output of information is usually much greater in length and more intelligible. Active engagement is critical !

5. Begin early on to collect objects/toys that can be used in your lessons with the student. These concrete objects will be used to actively engage the child in activities that enhance receptive and expressive language skills.

6. Allow the child to choose an object. Then, ask the child to tell all he/she can about the object. Encourage the child to analyze the size, texture, shape, composition, etc. This process of analyzing will enhance the forming of picture referents in the mind even when he/she is not able to verbally express them.

7. Continue to read aloud from books magazines and newspapers. Engage the child by asking questions about what was read or what will happen next. Then, when answering questions becomes too frustrating, provide two choices as possible answers. Have the student select the best answer. Yes/no questions can also be used.

8. If the child experiences difficulty with yes/no questions, the use of "silly questions" is a good way to promote the processing of auditory information. For example, ask, "Do chairs eat ?" "Do boys ear ?" Laugh about it and explain why it is silly. Have fun with these "silly questions".

9. "Audio-describe" TV shows and activities that are taking place in the classroom and at home. Talk about what you see, hear, smell, touch and taste.

10. Tape record sound clues, from field trips, vacations, the home and school environment, etc., that can be used as prompts to aid in the recall of information.

11. Keep a taped journal or log of experiences and events that the student can listen to throughout the years.

12. Over time, the facial and palatial muscles appear to lose their mobility and preciseness. Exercising the facial muscles to enhance the intelligibility of speech by singing songs, reciting poems and telling stories that use repetitive sounds and words.

13. Try singing a question to the student. Encourage the student to sing the answer. Compare intelligibility of responses.

14. A stress free environment will promote a more intelligible speech. Do not put pressure on the child to speak correctly or promptly.

15. Connected speech will be the least intelligible. Encourage two to three word utterances for more intelligible speech.

16. Coordinate speech work with physical therapy and occupational therapy activities.

17. Keep activities as "hands-on" as possible by using objects, role playing, interviews, skits, etc.

18. Encourage communication by talking together about situations with which the student is familiar, using gestures to enhance understanding, incorporating the use of signs when appropriate, and exercising facial muscles. Continue to provide "input" even when the child is not able to provide verbal "output".

Communication

Speech becomes difficult for the child and as it worsens it can become difficult to be understood. Occasionally having the child singsong words will help as singing requires a different thought/ muscle process as does speech. Eventually speech will be lost completely. Alternative communications need to be considered before that happens.

Communication is the most important area of instruction for the student with Batten Disease. Always teach with the long-term goal of enabling the student to communicate in the final stages of the disease. Being able to communicate will empower the student and give great comfort to the student in a time when many people underestimate the child's ability to understand and know his or her own wants and needs.

First, if your student still has verbal ability, tape record his or her voice expressing favorite sayings, requesting favorite objects and activities, greetings, and anything else important to the student. Establish an audiotaped library of these expressions for use in the future. Many augmentative communication devices are programmed with a real voice. Such devices, when they become necessary, can then be programmed with the student's own voice. Hearing his or her own voice when verbal skills are gone is a source of comfort and empowerment and motivates the student to access this mode of communication. If speech is already gone, use friends' and/or family members' voices to program the augmentative communication device.

Next, pair activities with objects that represent that activity. For example, a young girl loves to ride horses. Her mother paired this activity with a rein. Then, every time the girl was going to ride horses she would hold this rein on her way to this favorite activity. Once the objects are clearly established in the student's mind switch to a part of the object to represent the activity. In the above example, the part of the object would simply be a piece of a rein. These parts of objects can then be attached with Velcro to an augmentative device.

In this way, the student with visual impairment and limited verbal abilities can find the part of the object and press it to activate the augmentative device to request the activity (fig 2). Later, when the ability to differentiate the parts of objects becomes limited, return to the whole object to allow the student to continue to request desired activities, wants, and needs (fig 3). Objects and parts of objects are more concrete than textures or Braille and are easier for the student to continue to distinguish even when abilities to differentiate become limited. Also, teaching the objects early on in the process helps to establish the concept in the student's long-term memory. This is important because long-term memory appears stronger than short-term memory as the disease progresses.

The student with Batten Disease may feel out of control as he or she experiences the regression of skills. The student may communicate this feeling through aggression, swearing, or agitation. Providing structure in this student's day will assist the student in feeling more in control. Utilizing a calendar system for the student with Batten Disease is an excellent way to both provide structure and to practice the communication system of objects/parts of objects corresponding to activities.

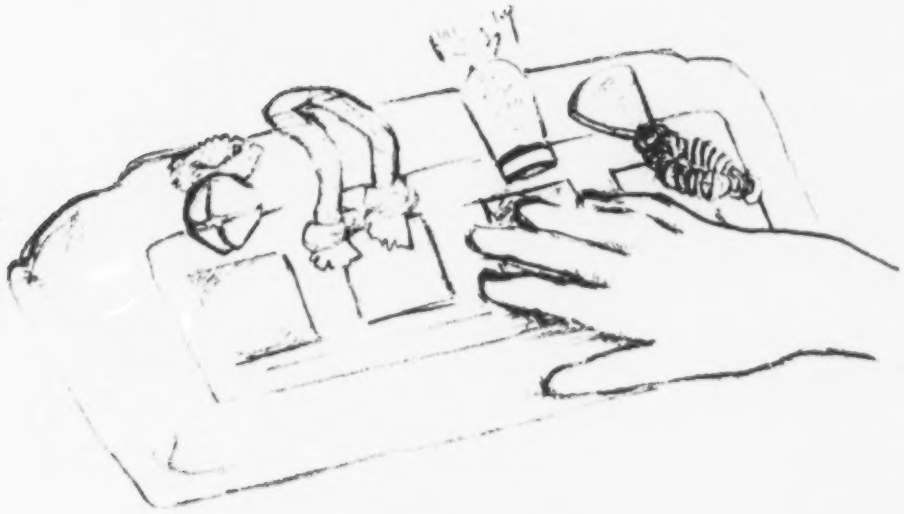

Figure 2. A four location augmentative device. The piece of rope on location #2 is a part of the rope that holds a swing. When location #2 is pressed the device says, "I'd like to swing, please."

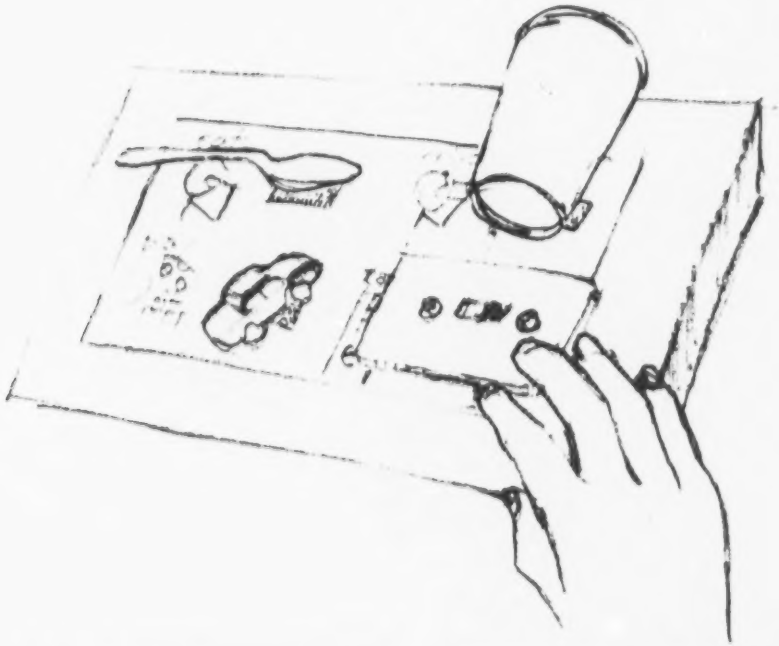

Figure 3. A four location augmentative device with whole objects attached with Velcro, representing eat, drink, play, and listen to music. The student is able to press the object to request the desired activity.

Emily, for instance, began with a system consisting of a series of eight boxes. Each box contained an object that corresponded to an activity planned for that day. Assisted by her teacher, Emily would open the first box, complete the activity, return the object to a "finished" box, and repeat the process with the next box. The number of boxes in the system decreased as time went on. However, the structure of the calendar system brought comfort to Emily. She knew what was going to happen in her day. When Emily became agitated, merely holding the calendar box would calm her and the agitation would vanish.

Daily Calendars. A routine or pattern is established when the calendar box objects are used in a set order time after time. This is recommended when beginning a calendar system to enable the student to mentally connect the object with the activity. When setting up a calendar system, it is recommended to run it consistently for a one month period before making any changes. This allows sufficient time to establish understanding of the objects or symbols and of the routine. At the end of one month, make any necessary changes and run for another month. Continue in this fashion until the student understands the function of the calendar box.

The next step in calendaring is to add objects or symbols that represent additional activities. The student will soon have more objects/activities than can be accomplished in one day. At this time, the student begins to make choices by choosing what activities he or she wants and in what order. Choices are made at the beginning of the day, so the student is actually planning his or her own day. As choices are made, the object or symbol goes into the box at the possible position. This choosing may take place in only one position of the calendar box to begin with, but the choice empowers the student, in any case. The teacher will still maintain whatever control is necessary by offering only choices that are possible for that day and only in the box position that allows for choices.

Example: Upon arrival to school, Emily is given six of her calendar box objects. The teacher then asks Emily what she wants to do first. Emily loves lunch, so she chooses the spoon that represents lunch. The teacher says, "Good choosing, Emily, but lunch isn't until 12:00. Let's put the spoon in box #3. What else would you like to do today?" This process continues until all four of Emily's boxes are filled. The teacher then says, "You planned a very good day. Let's get started. Please find box #1."

In the example, the teacher maintained the control she needed while still allowing Emily to make choices. Behavior problems are quickly reduced when empowering the student with some control over her day. Remember, though, the teacher is still in control over the offered choices.

Sometimes it is not possible to offer the student a choice. For instance, the physical therapist may be scheduled to come in and do range of motion with the student right before lunch. In this case, the teacher may present the student with two objects and ask him or her which object represents range of motion. Again, the student is actively engaged in the planning of his or her day.

A calendar box system that works well consists of two magazine holders attached together back-to-back (fig. 4). One side will hold four or five small boxes that are the basis of the calendar

system. Each box will have a number or dots on it designating its position. Use raised numbers, dots, or Braille numbers when using the system with a visually impaired student. The other side of this system is called the "finished box." Upon completion of an activity, the student places the corresponding object or symbol in the "finished box." This is an excellent way to identify the close of an activity.

A great deal of verbal communication between teacher and student should take place while interacting with the calendar box. In fact, the presence of the calendar box actually reminds the teacher to communicate with the student. All too often, children with disabilities have things done to them or for them. Co-active involvement with the calendar box will reduce the occurrence of this problem. Again, the student feels empowered when the calendar box is used appropriately.

Individualized Education Plan (IEP) Goals.

IEP goals can be written to directly involve the calendar box. Following are examples of IEP goals used to increase skills in basic communication and in muscular control.

Basic Communication:
" When presented with an activity/object, _____ will vocalize to show anticipation of the routine/activity 80% of the time over three consecutive data days".
" Throughout the day when _____ is presented with his calendar box, he will reach out and touch the object to indicate his desire to do the corresponding activity 75% of the time for three consecutive data days."
" Throughout the day when _____ is presented with her calendar box, she will reach and grasp the object to indicate her desire to do the corresponding activity 80% of the time for three consecutive data days."

Muscular Control:
" When presented with his calendar box and given verbal prompts, _____ will open his calendar boxes at 75% accuracy over three consecutive data days."

" When presented with a calendar box to _____'s left side and paired with an auditory cue, _____ will actively turn her head towards the calendar box (moving from right to left) for 75% of trials over three consecutive data days."

As you can see, calendar box goals are directly related with many goal areas needed by children with disabilities. The calendar box provides motivation to work on tiresome goals by empowering the student with choices. This may be the only empowerment that some students have. Also, the calendar box enables the teacher to provide organization and the stability of a routine to the life of his or her students.

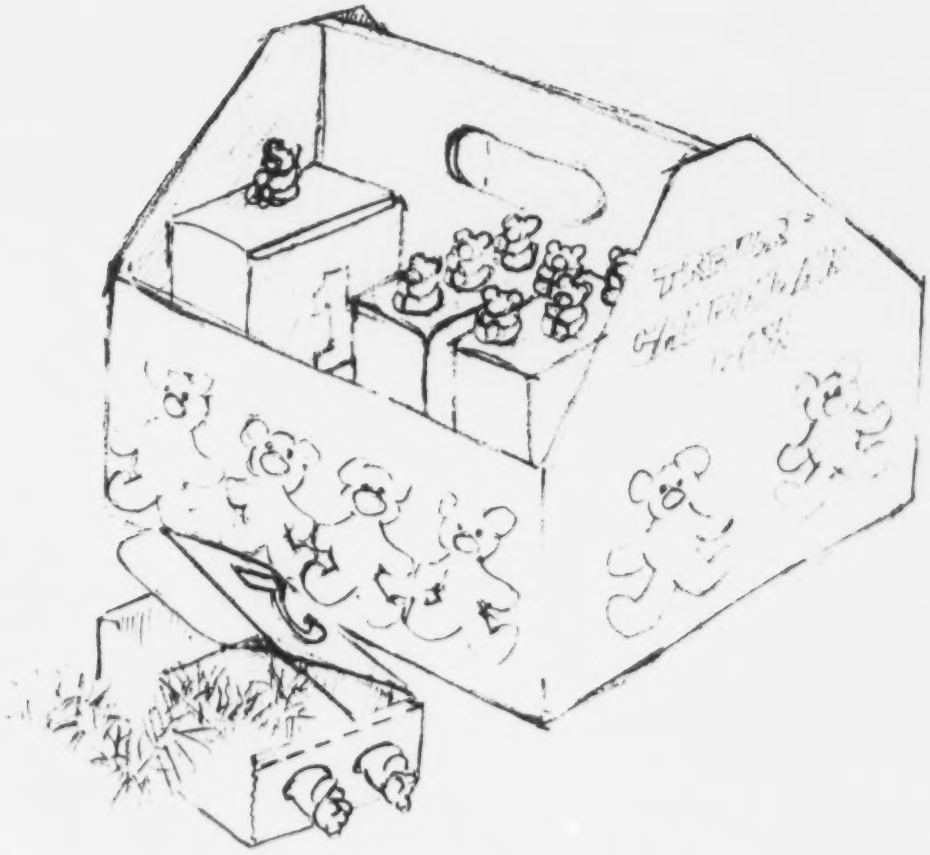

Figure 4. Trevor's calendar system contains four boxes. The tinsel in box #2 represents the sensory room. The backside of the calendar system is the "finished" box.

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Making choices is important to all students. Making choices is no less important to the student with Batten Disease. When verbal skills have declined, the student with Batten Disease can continue to make choices by choosing from the objects that correspond with activities. Even in the final stage of the disease, the student that has a communication system of objects can still make choices. This is accomplished by placing each of the student's hands on an object. Carefully observe for any movement, even a slight movement of the finger. This movement will designate the student's choice. What empowerment this choice-making ability will give the student!

Individuals with Disabilities Education Act 1997

Individualized Education Plan (IEP)

The 1997 Amendments expand the IEP content requirements significantly. First, present levels of performance (PLOPS) are expressed in objective, measurable terms to the extent possible. IDEA 1997 section 1414(d)(1)(A)(I) also mandates a statement of "how the child's disability affects the child's involvement and progress in the general curriculum."

Short-term objectives are series of achievable components of annual goals that build upon each other over the course of the school year. Appendix C to 34 CFR Part 300 defines annual goals as "statements that describe what a child with a disability can reasonably be expected to accomplish within a twelve-month period in the child's special education program." A statement of supplementary aids and services required must also be included in the IEP.

Section 1414(d)(4)(A)(ii) of the 1997 Amendments states that revisions to an IEP should address:

- Any lack of expected progress toward the annual goals
- The results of any re-evaluations conducted
- Information provided by the parents
- The child's anticipated needs; or
- Other matters

The following individuals must be present at all IEP meetings:

- Parents (must ensure they are invited and encouraged to attend)
- The child's teacher
- Regular education teacher (if the child may be participating in the regular education environment)
- Local agency representative (LEA)
- Individual(s) who can interpret the instructional implications of evaluation results (i.e. Psychologist, therapist, nurse, etc.)
- Representatives of other public agencies if child is 14 years old or older (34 CFR section 300.344 (c) requires the school to "invite" representatives likely to be involved with the student's planned transition services)

In addition, schools are required to permit the attendance of other individuals invited by the parents and are themselves permitted to invite other individuals. 34 CFR section 300.344(a)(5).

Parental Participation

To protect parents' rights in the IEP process, the school must provide notice, schedule for mutual convenience, and facilitate understanding of the process. 34 CFR section 300.345.

As directed by the Office of Special Education Programs, the content of the notice must include the purpose, time and location of the meeting, and who will be in attendance (sufficient to indicate by position only). 34 CFR section 300.345(b)(1).

According to Cordrey v. Euckert, 17 EHLR 104 (6th Cir. 1990), parents' unwillingness to participate will not invalidate the IEP if attempts to arrange a mutually convenient meeting and convince the parents to attend are documented. 34 CFR section 300.345(d).

Parents must also be involved in re-evaluation assessments and decisions.

Parental participation in placement decisions is mandated under the 1997. Amendments, regardless of whether those decisions are made by the IEP team or some other group. IDEA (as amended) section 1414(d).

Placement in Special Education

The team making placement decisions must include, of course, the parents. IDEA (as amended) section 1414(f). According to 34 CFR section 300.533(a), the team making placement decisions must:

- Draw upon information from a variety of sources
- Ensure that information is documented and carefully considered
- Ensure that the placement decision is made in conformity with the Least Restrictive Environment (LRE) rules

IDEA creates a preference for neighborhood schools. However, the court in Murray v. Montrose County School District RE-1J, 22 IDELR 558 (10th Cir. 1995) found that IDEA does not create a presumption in favor of neighborhood schools and therefore does not necessarily create a right to placement there. This is also evidenced by 34 CFR section 300.551(a) that requires school districts to ensure a continuum of alternative placements is available.

Center-based schools provide a centralization of services that increases the proficiency of personnel and maximizes the availability of staff. Cost also becomes an issue when serving students with low-incidence, high-cost disabilities. Regardless of the justification of and need for center-based schools, the IEP must now include an "explanation of the extent, if any, to which the child will not participate with nondisabled children in the regular class [and in the general education curriculum including extra-curricular and nonacademic activities]." IDEA (as amended) section 1414(d)(1)(A)(iv).

Related Services

Related services are developmental, corrective, or supportive services required to assist the child to benefit from special education.

The court in Irving Independent School District v. Tatro, 1983-84 EHLR 555:511 (1984) established a four-part test for distinguishing whether a particular health related service is covered under IDEA. Accordingly, the limitations include:

- The student must be IDEA-eligible
- The service must be necessary for the student to benefit from special education
- The student must need to be provided with the service during school hours
- The service can be performed by a nonphysician

School health care, as a related service, enables many medically fragile children to attend school. Concerning a paralyzed and ventilator-dependent adolescent in Cedar Rapids Community School District, 22 IDELR 278 (Iowa 1994), the court stated that the lack of stimulation the student now receives from teachers and peers would have a profound effect on the rest of his life. However, services needed at all times to sustain life are considered excluded medical services.

The administration of medication may be a related service.

Transportation to and from school is a related service. The duty to provide transportation as a related service begins at the curb or at the door depending on your particular district.

As assistive technology device may be a related service, special education, or a supple- mentary aid and service. IDEA (as amended) section 1414 (d)(3)(B)(v) requires the IEP team to "consider whether the child requires assistive technology devices and services."

Procedural Safeguards

Section 1415(b) of the 1997 Amendments states that the procedural safeguards include the following:

- Parents can examine all records, participate in identification, evaluation, placement, and decisions to provide a free, appropriate education (FAPE), and obtain an independent educational evaluation.
- Provision of surrogate parent if necessary.
- Written prior notice whenever the school A) proposes to initiate or change, or B) refuses to initiate or change the identification, evaluation, placement, or the provision of FAPE.
- Notice provided in native language of the parents.
- Opportunity for mediation
- Opportunity to present complaints.
- The procedures to provide notice when complaints are filed.
- Assistance in filing a complaint.

IDEA regulations define a "parent" as a parent, guardian, person acting as a parent of the child, or an appointed surrogate parent. The definition does not include the State if the child is a ward of the State. In addition, a foster parent may be a "parent" as a matter of state law.

Section 1415(c) of the 1997 Amendments specifies the contents of the prior written notice. The notice must include:

- Description of action proposed or refused
- Explanation of why
- Description of other options considered and reasons why rejected as basis for proposed action
- Description of any other relevant factors
- Statement of parents' rights and how to obtain a copy of the description of the procedural safeguards
- Sources for parents to contact to obtain assistance in understanding the provisions of this part.

In addition to prior written notice for identification, reevaluation, and placement, the school must also obtain parental informed consent prior to conducting a reevaluation. IDEA (as amended) section 1414(c)(3). To confirm that consent is informed, the parent must sign a written consent form that describes the action to be carried out. 34 CFR section 300.500(a)(2). Consent is voluntary and may be revoked at any time.
34 CFR section 300.500(a)(3).

Notes for the School Nurse

As a front line member of the medical staff, it is important that the school nurse be aware of a number of issues with regard to their students with Batten Disease.

The first of these are the seizures which may be presently experienced by the student. Indeed, if seizures are not currently manifesting themselves they do loom in the future, and they will be of several types. It is important to recognize that while medication should lessen the frequency of seizures, their eventual increasing occurrence is unavoidable. Contact with the student's neurologist, pediatrician, and parents may well be helpful in arriving at what has been typical for the child. Patterns of onset and frequency will vary from child to child.

Impairment of mobility will progress over time. A "bent knee, toe in" posture is typical for children with Batten Disease who are yet ambulatory.

Verbal issues which will likely be presented include increasing difficulty in articulation. Students might also be inclined to repeat sounds, syllables, and words over and over. A possible slowness in processing information, and delays in answering further indicate the need for accommodation.

One final notable tendency in children with Batten Disease is the maintenance of high motivation in the face of regressing skills. This growing gap may contribute to frustration for the student and, thus your support will be most beneficial.

Health Care Plan

Some of the health concerns that may require health care plans at school are the following:

· Seizures - Update protocols with increasing occurrence of seizures.
· Progressive Vision Loss - Orientation and mobility.
· Impaired Physical Mobility - Clumsiness progressing to wheelchair use and complete dependence; skin care; respiratory function; bowel function.
· Impaired Speech - Communication difficulties.
· Nutrition - Due to swallowing difficulties tube feeding may be necessary.
· Loss of Body Function - Incontinence; disturbance in self-concept.
· Dementia - Loss of control; memory loss; behavioral outbursts.
· Knowledge of Disease - Student, family, and caregivers.

Make Time Count!

You, as the teacher of a student with Batten Disease, can enhance the quality of life for your student now and in the future. What you do today can make a difference for the rest of your student's life. With knowledge of the disease and the importance of the instruction you provide, you can make time with your unique and special student count. The journey is just beginning for you. Make it a rewarding one for your student, for his or her family, and for yourself. The lessons you learn from your student with Batten Disease will enable you to become a better teacher to all students that you encounter in your teaching career. Good luck and God bless. Make it count!

The BDSRA values your experience and expertise. If you have any ideas, suggestions, or interventions that work, please let us know! Also, for further information contact:

Batten Disease Support and Research Association
2600 Parsons Ave., Columbus, Oh 43207
1-800-448-4570

Contact persons:

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